



# CLINICAL GUIDELINES

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## Pediatric Cardiac Imaging Policy

Version 20.0.2018  
Effective May 17, 2018



eviCore healthcare Clinical Decision Support Tool Diagnostic Strategies: This tool addresses common symptoms and symptom complexes. Imaging requests for individuals with atypical symptoms or clinical presentations that are not specifically addressed will require physician review. Consultation with the referring physician, specialist and/or individual's Primary Care Physician (PCP) may provide additional insight.

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<b>Procedure Codes Associated with Cardiac or PVD Imaging</b>	
<b>MRI/MRA</b>	<b>CPT®</b>
Cardiac MRI without contrast	75557
Cardiac MRI without contrast; with stress imaging	75559
Cardiac MRI without and with contrast	75561
Cardiac MRI without and with contrast; with stress imaging	75563
Unlisted MRI procedure (for radiation planning or surgical software)	76498
<b>CT</b>	<b>CPT®</b>
Heart CT with contrast for structure and morphology	75572
Heart CT with contrast for structure and morphology, for congenital heart disease	75573
Heart CT with contrast for coronary arteries & bypass grafts	75574
<b>CTA</b>	<b>CPT®</b>
CTA Abdominal Aorta with Bilateral Iliofemoral Runoff	75635
<b>Nuclear Medicine</b>	<b>CPT®</b>
PET Imaging; limited area (this code not used in pediatrics)	78811
PET Imaging; skull base to mid-thigh (this code not used in pediatrics)	78812
PET Imaging; whole body (this code not used in pediatrics)	78813
PET with concurrently acquired CT; limited area (this code rarely used in pediatrics)	78814
PET with concurrently acquired CT; skull base to mid-thigh	78815
PET with concurrently acquired CT; whole body	78816
PET Myocardial – Metabolic	78459
PET Myocardial Perfusion Imaging, Rest or Stress	78491
PET Myocardial Perfusion Imaging, Rest and Stress	78492
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SPECT Equilibrium Cardiac Radionuclide Angiography	78494
SPECT Equilibrium Multiple Cardiac Radionuclide Angiography	78496

Ultrasound	CPT®
Transthoracic echocardiography for congenital cardiac anomalies; complete	93303
Transthoracic echocardiography for congenital cardiac anomalies; follow-up study	93304
Echocardiography, transthoracic, real time with image documentation (2D), includes M-mode recording, when performed, complete, with spectral Doppler echocardiography, and with color flow Doppler echocardiography	93306
Echocardiography, transthoracic, real time with image documentation (2D), includes M-mode recording, when performed, complete, without spectral or color Doppler echocardiography	93307
Echocardiography, transthoracic, real time with image documentation (2D), includes M-mode recording, when performed, follow-up or limited study	93308
Doppler echocardiography, pulsed wave and/or continuous wave with spectral display (List separately in addition to codes for echocardiographic imaging); complete	93320
Doppler echocardiography, pulsed wave and/or continuous wave with spectral display (List separately in addition to codes for echocardiographic imaging); follow-up or limited study	93321
Doppler echocardiography color flow velocity mapping (List separately in addition to codes for echocardiographic imaging)	93325
Echocardiography, transesophageal, (TEE) real-time with image documentation (2D) (with or without M-mode recording); including probe placement, image acquisition, interpretation and report	93312
Echocardiography, transesophageal, (TEE) real-time with image documentation (2D) (with or without M-mode recording); placement of transesophageal probe only	93313
Echocardiography, transesophageal, (TEE) real-time with image documentation (2D) (with or without M-mode recording); image acquisition, interpretation & report only	93314
Transesophageal echocardiography (TEE) for congenital cardiac anomalies; including probe placement, image acquisition, interpretation and report	93315
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Transesophageal echocardiography (TEE) for monitoring purposes, including probe placement, real-time 2D image acquisition and interpretation leading to ongoing assessment of cardiac pumping function and to therapeutic measures on an immediate time basis	93318

**PEDCD-1: General Guidelines**

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### **PEDCD-1.1: Pediatric Cardiac Imaging Age Considerations**

Many conditions affecting the heart in the pediatric population are different diagnoses than those occurring in the adult population. For those diseases which occur in both pediatric and adult populations, minor differences may exist in management due to patient age, comorbidities, and differences in disease natural history between children and adults.

- Patients age < 18 years old should be imaged according to the Pediatric Cardiac Imaging Guidelines, and patients age ≥ 18 years should be imaged according to the Cardiac Imaging Guidelines, except where directed otherwise by a specific guideline section.

### **PEDCD-1.2: Pediatric Cardiac Imaging Appropriate Clinical Evaluation**

- A recent (within 60 days) face-to-face evaluation should be performed prior to considering advanced imaging, unless the patient is undergoing guideline-supported scheduled follow-up imaging evaluation. This evaluation should include:
  - ◆ a detailed history
  - ◆ physical examination
  - ◆ appropriate laboratory studies
- Unless otherwise stated in a specific guideline section, the use of advanced imaging to screen asymptomatic patients for disorders involving the heart is not supported.
- Advanced imaging of the heart should only be approved in patients who have documented active clinical signs or symptoms of disease involving the heart.
- Unless otherwise stated in a specific guideline section, repeat imaging studies of the heart are not necessary unless:
  - ◆ there is evidence for progression of disease
  - ◆ new onset of disease and/or documentation of how repeat imaging will affect patient management or treatment decisions.

### **PEDCD-1.3: Pediatric Cardiac Imaging Modality General Considerations**

- MRI
  - ◆ MRI and MRA studies are frequently indicated for evaluation of complex congenital heart defects not well visualized on echocardiography, and right ventricular disease.
  - ◆ Due to the length of time for image acquisition and the need for stillness, anesthesia is required for almost all infants and young children (age < 7 years), as well as older children with delays in development or maturity. In this patient population, MRI imaging sessions should be planned with a goal of avoiding a short-interval repeat anesthesia exposure due to insufficient information using the following considerations:
    - MRI should always be performed without and with contrast unless there is a specific contraindication to gadolinium use since the patient already has intravenous access for anesthesia.

- If multiple body areas are supported by eviCore guidelines for the clinical condition being evaluated, MRI of all necessary body areas should be obtained concurrently in the same anesthesia session.
- CT
  - ◆ CT is primarily used to evaluate the coronary and great vessels in congenital heart disease.
  - ◆ Coding considerations are listed in **PEDCD-10: CT Heart and Coronary Computed Tomography Angiography (CCTA- Other Indications)**
- Ultrasound
  - ◆ Echocardiography is the primary modality used to evaluate the anatomy and function of the pediatric heart, and is generally indicated before considering other imaging modalities.
  - ◆ Coding considerations are listed in **PEDCD-8: Echocardiography- Other Indications.**
- Nuclear Medicine
  - ◆ Multi Gated Acquisition (MUGA) studies (CPT® 78472, CPT® 78473, CPT® 78481, CPT® 78483, CPT® 78494, or CPT® 78496) are rarely performed in pediatrics, but can be approved for the following:
    - Certain pediatric oncology patients when echocardiography is insufficient: See: **PEDONC-1.2: Pediatric Oncology Imaging Appropriate Clinical Evaluations** for imaging guidelines.
    - Quantitation of left ventricular function when recent echocardiogram shows ejection fraction of < 50% and MUGA results will impact acute patient care decisions.
  - ◆ SPECT/CT fusion imaging involves SPECT (MPI) imaging and CT for optimizing location, accuracy, and attenuation correction combines functional and anatomic information.
    - There is currently no evidence-based data to formulate appropriateness criteria for SPECT/CT fusion imaging.
    - Combined use of nuclear imaging, including SPECT, along with diagnostic CT (fused SPECT/CT) is considered investigational.
  - ◆ Central C-V Hemodynamics (CPT® 78414) is not an imaging study, and is rarely performed.
    - If requested for a patient with congestive heart failure it may be approved after the requester is informed that this is NOT an imaging exam or MUGA examination.
    - CPT® 78414 should not be approved concurrently with any other 784xx CPT® code.
  - ◆ Cardiac Shunt Detection (CPT® 78428) is rarely performed in pediatrics but can be approved for the following:
    - Calculation of left and right ventricular ejection fractions
    - Assessment of wall motion
    - Quantitation of right to left shunts

- ◆ Infarct Avid Myocardial Imaging studies (CPT® 78466, CPT® 78468, and CPT® 78469) are obsolete examinations, which have been replaced by Cardiac MRI and are not supported by eviCore Imaging Guidelines.

The guidelines listed in this section for certain specific indications are not intended to be all-inclusive; clinical judgment remains paramount and variance from these guidelines may be appropriate and warranted for specific clinical situations.

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## **PEDCD-2: Congenital Heart Disease**

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## **PEDCD-2.1: Congenital Heart Disease General Considerations**

- Congenital heart disease accounts for the majority of cardiac problems occurring in the pediatric population. Patients may be diagnosed any time spanning prenatal evaluation to adolescence.
- There are a number of variables that influence the modality and timing of imaging patients with congenital heart disease, which results in a high degree of individuality in determining the schedule for imaging these patients, including:
  - ◆ patient age
  - ◆ physiologic effects of the defect
  - ◆ whether or not the defect has been repaired
  - ◆ rate of patient growth
  - ◆ stability of the defect on serial imaging
  - ◆ comorbid conditions
  - ◆ activity level

## **PEDCD-2.2: Congenital Heart Disease Echocardiography Coding**

- Any of the following echocardiography code combinations are appropriate for re-evaluation of patients with known congenital heart disease:
  - ◆ CPT® 93303, CPT® 93320, and CPT® 93325
  - ◆ CPT® 93304, CPT® 93321, and CPT® 93325
  - ◆ CPT® 93303
  - ◆ CPT® 93304
- CPT® 93306 is not indicated in the evaluation of known congenital heart disease.
- All requested CPT® combinations other than those listed in this section should be forwarded for Medical Director Review.

## **PEDCD-2.3: Congenital Heart Disease Modality Considerations**

- Echocardiography is the primary imaging modality used for monitoring congenital heart disease and is generally required before other imaging modalities are indicated unless otherwise indicated in a specific guideline section.
- Cardiac MRI either without contrast (CPT® 75557) or without and with contrast (CPT® 75561) is indicated for the following, when a recent echocardiogram is inconclusive:
  - ◆ CPT® 75565 is also indicated for patients with valvular disease or a need to evaluate blood flow through the chambers. These patients will usually have CPT® 93320 and CPT® 93325 performed with their echocardiography studies.
  - ◆ MRA Chest (CPT® 71555) may be added if the aorta or pulmonary artery needs to be visualized beyond the root, or if aortopulmonary collaterals, pulmonary veins, or systemic veins need to be visualized.
    - MRA Chest alone (CPT® 71555) should be performed if the patient cannot cooperate with full cardiac MRI exam.
- MRA Chest (CPT® 71555) is indicated for the following:

- ◆ Coarctation of the aorta (with or without Tetralogy of Fallot) with inconclusive recent echocardiography findings.
- CT imaging is indicated for the following:
  - ◆ Report CPT® 75574 for evaluating coronary artery anomalies
  - ◆ Report CPT® 75573 for congenital heart disease
  - ◆ Determination of extra-cardiac anatomy in patients with complex congenital heart disease
  - ◆ Pulmonary artery (PA) and Pulmonary vein (PV) assessment
  - ◆ Coarctation of the aorta or interruption of the aortic arch suspected on echocardiography.

### **PEDCD-2.4: Congenital Heart Disease Timing Considerations**

- Echocardiography is repeated frequently throughout a child's life, and the following intervals are within the standard of care and should be approved:
  - ◆ Patients age 0 to 2 years: every 3 months
  - ◆ Patients age 3 to 12 years: every 6 months
  - ◆ Patients age 13 years: every 12 months
  - ◆ Coarctation of the aorta: following repair, every 1 month until stable, then follow age-based guidelines.
  - ◆ Echocardiography is performed during the physician office visit, and these studies should not be denied because of lack of contact within 60 days.
  - ◆ Studies are often necessary more frequently than listed here, primarily due to changing symptomatology, or perioperative concerns. Requests not meeting these timing guidelines should be forwarded for Medical Director Review.
- The need for routine serial MRI or CT studies is uncommon, and these requests should be forwarded for Medical Director Review.

### ***References***

1. Bernstein D. Congenital Heart Disease. *Nelson Textbook of Pediatrics*. eds Kliegman RM, Stanton BF, St. Geme JW, et al. 19<sup>th</sup> edition 2011, pp. 1549-1610. Accessed November 30, 2017.
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## PEDCD-3: Heart Murmur

Heart murmurs are extremely common in pediatric patients. The thinner chest wall in children allows clearer auscultation of blood flowing through the chambers of the heart, which results in murmur on physical exam.

The majority of murmurs are innocent and do not require further evaluation. More than 30% of children will have an innocent murmur detected during physical examination. Innocent murmurs are typically systolic ejection murmurs with a vibratory or musical quality, and generally change in quality when the patient changes position.

Other types of murmurs are defined as pathologic and require additional evaluation, usually by a pediatric cardiologist. Echocardiography is indicated, and is performed as part of the office visit. When evaluating a patient with a murmur for the first time, it will not be known whether the patient has congenital heart disease or not. The cardiologist only submits charges for the procedure actually performed.

- The following echocardiography code combinations should be approved for evaluation of any pathologic murmur or any innocent murmur with associated cardiac signs or symptoms:
  - ◆ CPT® 93303, CPT® 93306, CPT® 93320, and CPT® 93325
  - ◆ CPT® 93303, CPT® 93306
  - ◆ CPT® 93306, CPT® 93320 and CPT® 93325 are included with CPT® 93306 and should not be approved separately.
- Repeat echocardiography is not indicated if the initial echocardiogram was normal and the murmur has not changed in quality.

### References

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## PEDCD-4: Chest Pain

Chest pain in pediatric patients is caused by a cardiac etiology in < 5% of cases, yet causes great anxiety for parents resulting in requests for testing.

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination, EKG, and appropriate laboratory studies should be performed prior to considering advanced imaging.
- Echocardiography is indicated for pediatric patients with chest pain and one or more of the following:
  - ◆ Exertional chest pain
  - ◆ Non-exertional chest pain with abnormal EKG
  - ◆ First-degree relative with sudden unexplained death or cardiomyopathy
  - ◆ Recent onset of fever
  - ◆ Recent illicit drug use
  - ◆ Other signs or symptoms of cardiovascular disease
- Echocardiography is performed as part of the office visit. When evaluating a patient for the first time, it will not be known whether the patient has congenital heart disease or not. The cardiologist only submits charges for the procedure actually performed.
- The following echocardiography code combinations should be approved for evaluation of chest pain:
  - ◆ CPT® 93303, CPT® 93306, CPT® 93320, and CPT® 93325
  - ◆ CPT® 93303, CPT® 93306
  - ◆ CPT® 93306
    - CPT® 93320 and CPT® 93325 are included with CPT® 93306 and should not be approved separately.
- Repeat echocardiography is not indicated if the initial echocardiogram is normal unless one of the following conditions is present:
  - ◆ Increased severity or change in quality of the chest pain
  - ◆ New signs or symptoms of cardiovascular disease other than pain
  - ◆ New abnormality on EKG

### References

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## PEDCD-5: Syncope

Syncope in pediatric patients is common, with up to 15% of patients experiencing at least one episode by age 21. Syncope is caused by neurocardiogenic syndrome (vasovagal syncope) in 75 to 80% of cases, which is a benign and self-limiting condition. Despite this, syncope causes great anxiety for parents resulting in requests for testing.

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination, EKG, and appropriate laboratory studies should be performed prior to considering advanced imaging.
- Echocardiography is not indicated for most patients with isolated syncope.
- Echocardiography is indicated for pediatric patients with syncope and one or more of the following:
  - ◆ Exertional syncope
  - ◆ Unexplained post-exertional syncope
  - ◆ Abnormal EKG
  - ◆ First-degree relative with any of the following before age 50:
    - Sudden cardiac arrest or death
    - Pacemaker or implantable defibrillator placement
  - ◆ First-degree relative with cardiomyopathy
  - ◆ Known congenital heart disease
  - ◆ History of Kawasaki disease
  - ◆ Pathologic murmur, irregular rhythm, gallop, or click on physical examination
- Echocardiography is performed as part of the office visit. When evaluating a patient for the first time, it will not be known whether the patient has congenital heart disease or not. The cardiologist only submits charges for the procedure actually performed.
- The following echocardiography code combinations should be approved for evaluation of chest pain:
  - ◆ CPT® 93303, CPT® 93306, CPT® 93320, and CPT® 93325
  - ◆ CPT® 93303, CPT® 93306
  - ◆ CPT® 93306
    - CPT® 93320 and CPT® 93325 are included with CPT® 93306 and should not be approved separately.
- Repeat echocardiography is not indicated if the initial echocardiogram is normal unless one of the following conditions is present:
  - ◆ Increased severity or change in quality of the syncope
  - ◆ New signs or symptoms of cardiovascular disease other than syncope
  - ◆ New abnormality on EKG

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1. Bernstein D. Evaluation of the Cardiovascular System, *Nelson Textbook of Pediatrics*. eds Kliegman RM, Stanton BF, St. Geme JW, et al. 19<sup>th</sup> edition 2011, pp. 1529-1549.
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## PEDCD-6: Kawasaki Disease

Kawasaki disease (KD) is the leading cause of acquired pediatric cardiac disease in the developed world. It is an acute febrile illness characterized by a medium vessel vasculitis, which predominantly affects the coronary arteries.

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination, and appropriate laboratory studies should be performed prior to considering advanced imaging.  
Patients who lack full clinical features of classic KD are often evaluated for incomplete KD. If coronary artery abnormalities are detected, the diagnosis of KD is considered confirmed in most cases
- If Kawasaki disease is strongly suspected, treatment will begin even before cardiac evaluation, since early treatment is associated with a lower risk for coronary aneurysm development.
- Echocardiography (CPT® 93306) is indicated for all patients with strongly suspected or known Kawasaki disease
  - ◆ Echocardiography is recommended at the time of diagnosis, 1 to 2 weeks later, and 6 weeks from diagnosis.
  - ◆ Patients with recurrent or persistent fever or worsening cardiac symptoms should have echocardiogram repeated.
  - ◆ Patients with no coronary abnormalities on 6 week study should have a repeat echocardiogram 1 year from diagnosis.
  - ◆ Patients with coronary abnormalities will require more frequent echocardiograms based on severity of disease and need for medical management. These requests should be forwarded for Medical Director Review.
- Coronary CTA (CPT® 75574), Cardiac MRI without contrast (CPT® 75557), Cardiac MRI without and with contrast (CPT® 75561), or MRA Chest (CPT® 71555) is indicated for evaluation of inconclusive echocardiogram findings, or for large coronary aneurysms.
- Screening of other body areas for aneurysms is not routinely indicated in Kawasaki disease, but MRA or CTA (contrast as requested) of the affected body area can be approved for evaluation of signs or symptoms suggesting aneurysm development.

### References

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## PEDCD-7: Pediatric Pulmonary Hypertension

Pulmonary hypertension in children can be caused by cardiac, pulmonary, or systemic diseases, and idiopathic disease occurs as well.

- A recent (within 60 days) face-to-face evaluation including a detailed history, physical examination, and appropriate laboratory studies should be performed prior to considering advanced imaging.
- If pulmonary hypertension is suspected, initial evaluation should consist of chest x-ray, EKG, and echocardiography (CPT® 93303, CPT® 93306, CPT® 93320, and CPT® 93325, see: **PEDCD-8.1: Transthoracic Echocardiography (TTE) Coding** for echocardiography coding considerations).
- Repeat echocardiography using pulmonary hypertension-specific protocols is indicated every 4 to 6 months for all pediatric patients with pulmonary hypertension.
  - ◆ Echocardiography is indicated at any time for new or worsening symptoms or to evaluate a recent change in therapy.
- CTA Chest (CPT® 71275) is indicated for initial evaluation of all pediatric patients with pulmonary hypertension to evaluate for pulmonary vascular or interstitial disease or other intrathoracic causes.
- Cardiac MRI without contrast (CPT® 75557) or without and with contrast (CPT® 75561) is indicated for evaluation of inconclusive echocardiogram findings, or for monitoring right ventricular function during follow-up.
- Screening of other body areas for aneurysms is not routinely indicated, but MRA or CTA (contrast as requested) of the affected body area can be approved for evaluation of signs or symptoms suggesting aneurysm development.

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**PEDCD-8: Echocardiography-Other Indications**

<b>PEDCD-8.1: Transthoracic Echocardiography (TTE) Coding</b>	<b>19</b>
<b>PEDCD-8.2: Initial Transthoracic Echocardiography (TTE) Indications</b>	<b>19</b>
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## **PEDCD-8.1: Transthoracic Echocardiography (TTE) Coding**

- CPT® codes for echocardiography are listed in **PEDCD-1: General Guidelines**
- The most commonly performed study is a complete transthoracic echocardiogram with spectral and color flow Doppler (CPT® 93306).
  - ◆ CPT® 93306 includes CPT® 93320 and CPT® 93325, so those codes should **not** be approved along with CPT® 93306.
  - ◆ Doppler codes (CPT® 93320, CPT® 93321, and CPT® 93325) are add-on codes and are assigned in addition to code for the primary procedure, and should not be approved alone.
- For a 2D transthoracic echocardiogram without Doppler, report CPT® 93307.
- Limited transthoracic echocardiogram should be billed if the report does not “evaluate or document the attempt to evaluate” all of the required structures.
  - ◆ A limited transthoracic echocardiogram is reported with CPT® 93308.
  - ◆ Unlike CPT® 93306, the Doppler CPT® 93321 and CPT® 93325 are not included with CPT® 93308. CPT® 93321 (not CPT® 93320) should be reported with CPT® 93308 if Doppler is included in the study. CPT® 93325 should also be reported with CPT® 93308 if color flow Doppler is included in the study.
  - ◆ For patients with congenital heart disease, a limited transthoracic echocardiogram is reported with CPT® 93304, +/- CPT® 93321 and CPT® 93325.
  - ◆ **NOTE:** Providers performing an initial echo on a pediatric patient will not know what procedure codes they will be reporting until the initial study is completed.
    - If congenital heart disease is found on the initial echo, a complete echo is reported with codes CPT® 93303, CPT® 93320, and CPT® 93325 because CPT® 93303 does NOT include Doppler and color flow mapping.
    - If no congenital issue is discovered, then CPT® 93306 is reported alone and includes 2-D, Doppler and color flow mapping.
    - Since providers may not know the appropriate code/s that will be reported at the time of the pre-authorization request, they may request multiple codes.
    - The following echocardiography code combinations should be approved for any initial echocardiogram:
      - CPT® 93303, CPT® 93306, CPT® 93320, and CPT® 93325
      - CPT® 93303, CPT® 93306
      - CPT® 93306
        - CPT® 93320 and CPT® 93325 are included with CPT® 93306 and should not be approved separately.
    - Depending upon individual health plan payer contracts, post-service audits may be completed to ensure proper claims submission.

## **PEDCD-8.2: Initial Transthoracic Echocardiography (TTE) Indications**

- In addition to indications listed in previous guideline sections, initial TTE evaluation is indicated for any of the following:
  - ◆ Any signs/symptoms that are possibly cardiac in nature, including (but not limited to) central cyanosis, dyspnea, edema, poor peripheral pulses, feeding difficulty, decreased urine output, hepatomegaly, or desaturation on pulse oximetry.

- ◆ Abnormal EKG or cardiac biomarkers
- ◆ Abnormal chest x-ray suggesting cardiovascular disease
- ◆ Palpitations and one of the following:
  - Abnormal EKG
  - First-degree relative with any of the following before age 50:
    - Sudden cardiac arrest or death
    - Pacemaker or implantable defibrillator placement
  - First-degree relative with cardiomyopathy
- ◆ Supraventricular Tachycardia (SVT), Ventricular Tachycardia, or Premature Ventricular Contractions (PVCs)
- ◆ Known or suspected valvular dysfunction
- ◆ Persistent systemic hypertension
- ◆ Obesity (BMI > 30) with additional cardiovascular risk factors
- ◆ Stroke
- ◆ Renal failure
- ◆ Preoperative evaluation of patients with chest wall deformities or scoliosis
- ◆ Known or suspected vascular ring
- ◆ Planned administration of cardiotoxic chemotherapy
  - Generally anthracyclines (doxorubicin, daunorubicin, mitoxantrone, idarubicin, epirubicin)
- ◆ Planned radiation therapy involving heart muscle or hematopoietic stem cell transplant
- ◆ Sickle cell disease or other hemoglobinopathy causing chronic anemia
- ◆ Known or suspected vasculitis, acute rheumatic fever, or other systemic autoimmune disease
- ◆ Muscular dystrophy
- ◆ Metabolic, mitochondrial, and storage disorders
- ◆ Abnormalities of cardiac or other viscera situs
- ◆ Signs, symptoms, or blood culture suggestive of endocarditis
- ◆ Known or suspected mass lesion involving heart or great vessels
- ◆ Known or suspected clot in atrium or ventricle
- ◆ Known or suspected pulmonary hypertension
- ◆ Known or suspected pericardial effusion
- ◆ Complications during prenatal development:
  - Known or suspected cardiovascular abnormality on fetal echocardiogram
  - Maternal phenylketonuria (PKU)
  - Maternal diabetes with no fetal echo
  - Maternal teratogen exposure
  - Maternal infection during pregnancy with potential cardiac sequelae
- ◆ Genetic abnormality known to be associated with cardiovascular disease
- ◆ First-degree relative family history of:
  - Unexplained sudden death before age 50
  - Hypertrophic cardiomyopathy
  - Non-ischemic dilated cardiomyopathy
  - Genetic abnormality known to be associated with cardiovascular disease
  - Congenital left-sided heart lesion

- Heritable pulmonary arterial hypertension

### **PEDCD-8.3: Repeat Transthoracic Echocardiography Indications**

Repeat echocardiograms are not necessary for most patients with clinically stable syndromes.

- In addition to indications listed in previous guideline sections, repeat TTE evaluation is indicated for any of the following:
  - ◆ New or worsening symptoms in a patient with known cardiac disease, previously normal echocardiogram with one of the following:
    - New or worsening cardiac symptoms
    - New EKG abnormality
    - Newly recognized family history suggestive of heritable heart disease
  - ◆ Every 12 months for patients age 12 to 18 years with first-degree family history of hypertrophic cardiomyopathy.
  - ◆ Every 12 months for patients receiving active therapy for ventricular hypertrophy, valvular dysfunction, cardiomyopathy.
    - One time repeat TTE can be approved at 6 months to assess response to a change in therapy.
  - ◆ Every 12 months for patients with chronic pericardial effusions
  - ◆ Every 12 months for sickle cell disease or other hemoglobinopathy causing chronic anemia and one of the following:
    - High risk genotype (Hgb SS or Sβ<sup>0</sup>, severe thalassemia, etc.)
    - History of acute chest syndrome or intrinsic lung disease
    - History of stroke
    - Receiving chronic transfusion therapy
  - ◆ As needed for monitoring cardiotoxicity during chemotherapy administration
  - ◆ After completion of chemotherapy and/or radiation therapy. See **PEDONC-19.2: Cardiotoxicity and Echocardiography** for imaging guidelines.

### **PEDCD-8.4: Transesophageal Echocardiography (TEE)**

- Transesophageal echocardiography imaging indications in pediatric patients are identical to those for adult patients. See **CD-2.5: Transesophageal Echocardiography (TEE)** in the Cardiac Imaging Guidelines.

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**PEDCD-9: Cardiac MRI-Other Indications**

<b>PEDCD-9.1: General Guidelines</b>	<b>24</b>
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## **PEDCD-9.1: General Guidelines**

Requests for cardiac MRI that contain only one CPT® code can be completed by the Nurse Reviewer. If the request contains more than one cardiac/chest MRI CPT® code, it should be forwarded for Medical Director Review.

- MRA of the coronary arteries is comparatively less accurate than CCTA or invasive coronary angiography in evaluating coronary disease and is considered investigational at this time.

## **PEDCD-9.2: Cardiac MRI - Coding**

<b>Cardiac Imaging Procedure Codes</b>	
<b>Cardiac MRI</b>	<b>CPT®</b>
Cardiac magnetic resonance imaging for morphology and function without contrast.	75557
Cardiac magnetic resonance imaging for morphology and function without and with contrast and further sequences.	75561
Cardiac magnetic resonance imaging for morphology and function without contrast; with stress imaging (rarely used in pediatrics).	75559
Cardiac magnetic resonance imaging for morphology and function without and with contrast and further sequences; with stress imaging (rarely used in pediatrics).	75563
Cardiac magnetic resonance imaging for velocity flow mapping (List separately in addition to code for primary procedure).	+75565

- Only one procedure code from the set: CPT® 75557, CPT® 75559, CPT® 75561, and CPT® 75563 should be reported per session.
- Only one flow velocity measurement (CPT® +75565) should be reported per session.

## **PEDCD-9.3: Indications for Cardiac MRI**

- In addition to indications listed in previous guideline sections, Cardiac MRI evaluation is indicated for any of the following, when a recent TTE is inconclusive:
  - ◆ Assessment of global ventricular function and mass if a specific clinical question is left unanswered by recent TTE and the MRI results will affect management of the patient's condition.
  - ◆ Clinical suspicion of arrhythmogenic right ventricular dysplasia (ARVD) or arrhythmogenic cardiomyopathy (ARVC).
    - MRI without contrast (CPT® 75557) is considered the optimal test for this disorder.
  - ◆ For pericardial disease (including constrictive pericarditis, restrictive pericarditis, and perimyocarditis), MRI should not be utilized to diagnose pericarditis but only to answer the question regarding possible constriction or restriction suggested clinically or by other techniques (TTE, etc.).



- MRI without and with contrast (CPT® 75561) is considered the optimal test for this disorder.
- ◆ Evaluate cardiac tumor or mass
  - MRI without and with contrast (CPT® 75561) is considered the optimal test for this disorder.
- ◆ Evaluate anomalous coronary artery
  - MRI without and with contrast (CPT® 75561) or CCTA (CPT® 75574) is considered the optimal test for this disorder.
- ◆ For Fabry's disease, late enhancement MRI may predict the effect of enzyme replacement therapy on myocardial changes that occur with this disease.
  - MRI without and with contrast (CPT® 75561) is considered the optimal test for this disorder.
- ◆ For Cardiomyopathy, Cardiac MRI can be performed to evaluate patients with congenital cardiomyopathy (muscular dystrophy, glycogen storage disease, fatty acid oxidation disorders, mitochondrial disorders, etc.) or unexplained cases of cardiomyopathy in order to characterize the myocardium.
- ◆ Cardiac stress perfusion study (CPT® 75559 or CPT® 75563) can be considered on a case by case basis for patients with anomalous coronary artery, Kawasaki disease, or other disorder with the potential for coronary ischemia who cannot undergo other forms of stress testing (ETT, MPI, etc.).
- ◆ Assessment of cardiac iron overload in hemochromatosis (either CPT® 75557 or CPT® 71550, T2\* MRI, contrast not necessary).
  - Screening imaging may be approved every 12 months
  - Imaging may be approved every 3 months for treatment response in patients receiving active treatment (chelation +/- phlebotomy)
  - Frequently performed along with MRI Abdomen (CPT® 74181) to assess liver iron deposition. See **PEDAB-18.2: Transfusion-Associated (Secondary) Hemochromatosis** for additional imaging guidelines.

#### **PEDCD-9.4: Aortic Root and Proximal Ascending Aorta**

- For screening due to family history of aortic aneurysm or dissection, see: **CH-29: Thoracic Aorta** in the adult Chest Imaging Guidelines.
- For patients who have both cardiac and ascending aorta abnormalities, the following studies are indicated:
  - ◆ Cardiac MRI (CPT® 75557 or CPT® 75561) for patients with abnormalities involving the aortic root and/or proximal ascending aorta.
  - ◆ If the distal ascending aorta is involved, MRI Chest (CPT® 71552) or MRA Chest (CPT® 71555) is also indicated.
- For patients with aortic abnormalities without cardiac abnormalities, any of the following studies is indicated:
  - ◆ MRI Chest (CPT® 71552)
  - ◆ MRA Chest (CPT® 71555)

## **PEDCD-9.5: Evaluation of Pericardial Effusion or Diagnosis of Pericardial Tamponade**

Echocardiogram is the initial imaging study of choice to evaluate pericardial effusions or diagnose pericardial tamponade. However, contrast enhanced cardiac MRI is useful for evaluating pericarditis, neoplastic effusion, tamponade or myocardial infiltration if a specific clinical question is left unanswered by another recent imaging study and the answer to the clinical question will affect management of the patient's clinical condition.

Cancers that can metastasize to the pericardium or myocardium and can cause a malignant effusion include lung, breast, renal cell, lymphoma and melanoma.

### ***References***

1. Bernstein D. Evaluation of the cardiovascular system, *Nelson Textbook of Pediatrics*. eds Kliegman RM, Stanton BF, St. Geme JW, et al. 19<sup>th</sup> edition 2011, pp. 1529-1549.
2. Atweh LA, Orth RC, Guillerman P, et al. MR imaging of children and young adults with classic findings of osteonecrosis on unenhanced MR imaging: do contrast-enhanced sequences help? *Pediatr Radiol*. 2013 Nov;43(11):1502-1506. Accessed December 27, 2017. <https://link.springer.com/article/10.1007%2Fs00247-013-2714-1>.

## **PEDCD-10: CT Heart and Coronary Computed Tomography Angiography (CCTA)-Other Indications**

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<b>PEDCD-10.5: Radiation Dose</b>	<b>29</b>

## **PEDCD-10.1: General Considerations**

Most payers require cardiac CT studies to be performed on a 64-slice CT scanner.

Metal artifact reduces the accuracy of CCTA. Devices that can cause this issue include, but are not limited to, surgical clips, pacemaker devices, defibrillator devices and tissue expanders.

Cardiac testing that does not involve exposure to ionizing radiation should be strongly considered.

Multi-slice CT is associated with a non-negligible risk for cancer, especially in women and younger patients.

### ***Practice Note***

<b>Relative Contraindications to CCTA Include:</b>	
1.	Irregular heart rhythms (e.g. atrial fibrillation/flutter, frequent irregular premature ventricular contractions or premature atrial contractions, and high grade heart block)
2.	Very obese patients (body mass index > 40 kg/m <sup>2</sup> )
3.	Elevated calcium score: CCTA should not be performed if there is extensive coronary calcification (calcium score >1000).
4.	Renal insufficiency
5.	Inability to follow breath holding instructions
6.	Heart rate over 75 beats per minute
7.	Allergy to iodine contrast material

## **PEDCD-10.2: Anomalous Coronary Artery**

- Evaluating coronary artery anomalies and other complex congenital heart disease of cardiac chambers or great vessels is an appropriate indication for CCTA.
  - ◆ Report CPT® 75574 for evaluating coronary artery anomalies
  - ◆ Report CPT® 75573 for congenital heart disease
  - ◆ Can add CPT® 71275 (chest CTA) to evaluate great vessels
  - ◆ In cases of anomalous pulmonary venous return, can add CT abdomen and pelvis with contrast (CPT® 74177).

Aortic root echocardiography or Cardiac MRI can be approved in lieu of CCTA to avoid radiation exposure.

**The use of CCTA to rule out anomalous coronary artery should be limited to one of the following:**

- Patients who need to have an anomalous coronary artery mapped prior to an invasive procedure.
- Patients who have not had a previous imaging study that clearly demonstrates an anomalous coronary artery.
- Previous imaging study shows the anomalous artery to be patent.

- Patients with a history that includes one or more of the indications in **PEDCD-10.3: Indications for CCTA**

### **PEDCD-10.3: Indications for CCTA (CPT® 75574)**

- In addition to indications listed in previous guideline sections, CCTA is indicated for any of the following, when a recent TTE and/or MRI is inconclusive:
  - ◆ Persistent exertional chest pain and normal stress test
  - ◆ Full sibling(s) with history of sudden death syndrome before age 30 or with documented anomalous coronary artery
  - ◆ Resuscitated sudden death and contraindication to conventional coronary angiography
  - ◆ Unexplained new onset of heart failure if CCTA will replace conventional invasive coronary angiography
  - ◆ Documented ventricular tachycardia (6 beat runs or greater) if CCTA will replace conventional invasive coronary angiography
  - ◆ Equivocal coronary artery anatomy on conventional cardiac catheterization
  - ◆ In infants: otherwise unexplained dyspnea, tachypnea, wheezing, episodic pallor, irritability, sweating, poor feeding, and/or failure to thrive
    - The presence of other congenital heart disease is not a separate indication for CCTA to rule out anomalous coronary artery
  - ◆ Evaluation of the arterial supply and venous drainage in children with bronchopulmonary sequestration

### **PEDCD-10.4: Indications for Cardiac CT (CPT® 75572)**

- In addition to indications listed in previous guideline sections, CCTA is indicated for any of the following, when a recent TTE and/or MRI is inconclusive:
  - ◆ Cardiac or pericardial mass
  - ◆ Pericarditis
  - ◆ Complications of cardiac surgery or evaluation of post-operative anatomy
  - ◆ Cardiac thrombus in patients with technically limited TTE, TEE, or MRI
  - ◆ Clinical suspicion of arrhythmogenic right ventricular dysplasia (ARVD) or arrhythmogenic cardiomyopathy (ARVC)
  - ◆ Native aortic abnormalities if echocardiogram is indeterminate

### **PEDCD-10.5: Radiation Dose**

- Radiation dosage for CCTA varies by facility and the particular protocol used. The American College of Radiology Clinical Statement on Noninvasive Cardiac Imaging states that “as a general rule a multi-detector CT encompassing the heart should not result in an effective dose of greater than 12 mSv.”\*
- 64-slice CT scanners can deliver a radiation dose from 15 to 25 mSv (especially in women due to breast tissue density).
- Sophisticated gating and other techniques can reduce the radiation dose of cardiac CT studies to less than 5 mSv. Application of these techniques is increasing nationwide.\*

- Dual source scanners decrease radiation exposure by approximately one third.
- Conventional coronary angiography typically delivers a radiation dose of 3 to 6 mSv.\*
- Newer imaging technologies will allow for reduced radiation exposure.

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